



Double Outlet Right Ventricle with Discordant Atrioventricular Connections in a Newborn: Case Report and Brief Overview of the Literature

Yenidoğanda Diskordan Atriyoventriküler Bağlantılı Çift Çıkışlı Sağ Ventrikül: Olgu Sunumu Ve Literatüre Kısa Bir Bakış

Discordant Atrioventricular Connections

Ahmet Sert¹, Ebru Aypar¹, Zeynel Gökmen², Mehmet Öç³, Dursun Odabaş¹
¹Department of Pediatric Cardiology, Konya Training and Research Hospital,
²Department of Neonatology, Konya Training and Research Hospital,
³Department of Cardiovascular Surgery, Selcuk University Hospital, Konya, Turkey

Özet

Çift çıkışlı sağ ventrikül bütün doğumsal kalp defektlerinin nadir bir kardiyak malformasyondur. Anormal ventriküloarteryal bağlantıların heterojen bir grubudur. Her iki büyük arter (pulmoner arter ve aorta) esas olarak morfolojik sağ ventrikülden çıkmaktadır. Diskordan atriyoventriküler bağlantılı çift çıkışlı sağ ventrikül ve hipoplazik pulmoner arterleri olan bir yenidoğanı sunmaktayız. Ekokardiyografi ile tanı konulan nadir karmaşık bir kardiyak malformasyonu bildirdik. Ekokardiyografi ile segmental yaklaşım ve kardiyak segmentlerin ardışık analizi karmaşık kardiyak malformasyonların tanısında ipucudur.

Anahtar Kelimeler

Çift Çıkışlı Sağ Ventrikül; Ekokardiyografi; Yenidoğan

Abstract

Double outlet right ventricle is a rare cardiac malformation of all congenital cardiac defects. It is a heterogeneous group of abnormal ventriculoarterial connections. Both great arteries (pulmonary artery and aorta) arise primarily from the morphological right ventricle. Double outlet right ventricle with discordant atrioventricular connections have been reported rarely in the literature. We report a newborn with double outlet right ventricle with discordant atrioventricular connections and hypoplastic pulmonary arteries. We reported a rare complex cardiac malformation diagnosed by echocardiography. Segmental approach and sequential analysis of cardiac segments by echocardiography are the clue for diagnosis of complex cardiac malformations.

Keywords

Double Outlet Right Ventricle; Echocardiography; Newborn

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Corresponding Author: Ahmet Sert, Department of Pediatric Cardiology, Konya Training and Research Hospital, 42080, Konya, Turkey.

T.: +90 3323236709 F.: +90 3323236723 E-Mail: ahmetsert2@hotmail.com

Introduction

Double outlet right ventricle is a heterogeneous group of abnormal ventriculoarterial connections where, by definition, both great arteries (pulmonary artery and aorta) arise primarily from the morphologically right ventricle [1]. It is a rare cardiac malformation, accounting for fewer than 1% of all congenital cardiac defects, and occurring in 1/10,000 live births [2,3].

The anatomical types are classified according to the following: a) the relationship between the ventricular septal defect (ventricular septal defect and the blood vessels); b) the position of the great arteries in relation to each other and c) the presence of additional malformations. According to the relationship between the ventricular septal defect and the blood vessels, double outlet right ventricle can be classified as follows: a) double outlet right ventricle with subaortic ventricular septal defect; b) double outlet right ventricle with subpulmonary ventricular septal defect; c) double outlet right ventricle with doubly-committed ventricular septal defect; and d) double outlet right ventricle with non-committed ventricular septal defect [2].

Double outlet right ventricle with discordant atrioventricular connections has been described rarely in the literature [2]. We report a newborn with double outlet right ventricle with discordant atrioventricular connections and hypoplastic pulmonary arteries.

Case Report

A 3100 grams male infant was born via spontaneous vaginal delivery at 38 weeks gestation after an uneventful pregnancy to a primigravida mother. Apgar scores were 7 at 1 and 5 minutes, respectively. He was the first sibling of nonconsanguineous parents. Shortly after birth, he was transferred to neonatal intensive care unit for dyspnea and cyanosis. On admission, physical examination revealed: weight: 3010 grams (25-50 p); height 46 cm (<3 p); head circumference: 33.5 cm (3-10 p); pulse rate: 124/min; respiratory rate: 64/min; blood pressure: 57/29 mmHg and temperature: 36.2Co. His general state was poor. He had severe cyanosis, tachypnea and retraction of the breath muscles. His heart auscultation revealed a grade 2 to 3/6 systolic murmur best heard on the upper left sternal border. His pulses were symmetrical and had normal amplitude. Other examinations were normal. Chest x-ray revealed mesocardia and decreased pulmonary vascular markings. The electrocardiogram showed superior QRS axis, and right ventricular hypertrophy. Echocardiography showed situs solitus, double outlet right ventricle with discordant atrioventricular connections, and large (10 mm) doubly-committed ventricular septal defect. The aorta and the pulmonary artery arose from the right ventricle, great arteries lied side by side, and the aortic and pulmonary valves were at the same level. Right atrium emptied through the mitral valve into the left ventricle, and left atrium emptied through the tricuspid valve into the right ventricle. The pulmonary annulus, main pulmonary artery and pulmonary artery branches were hypoplastic (3 mm). Infundibular and valvular pulmonary subvalvular stenosis with a pressure gradient of 45 mmHg, a 4 mm ostium secundum type atrial septal defect and normal pulmonary venous drainage were noted. Systemic veins connected to the right-sided morphologic right atrium (Figure 1, 2, 3). Abdominal ultrasonography was normal.

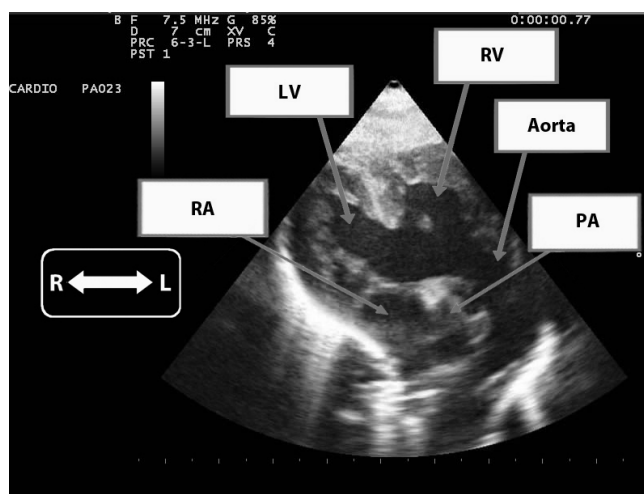


Figure 1. Transthoracic echocardiography subcostal long-axis views. Two dimensional view demonstrating double outlet right ventricle. Both aorta and pulmonary artery arise from the right ventricle. Pulmonary and aortic valves are at the same level. Note large doubly-committed ventricular septal defect between left ventricle and right ventricle.

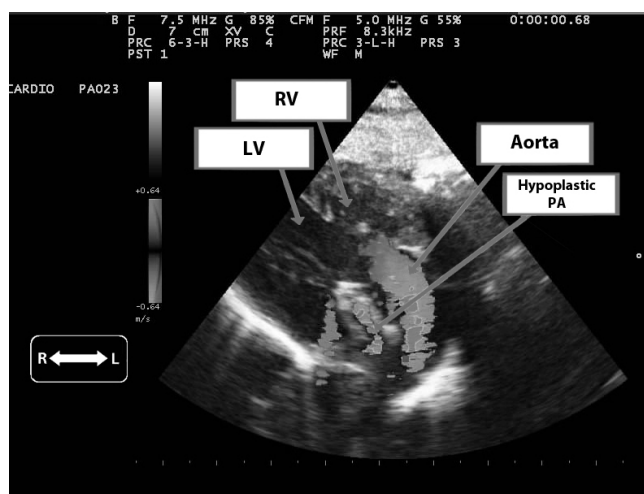


Figure 2. Transthoracic echocardiography subcostal long-axis views. Color Doppler view showing mosaic flow in hypoplastic pulmonary artery.

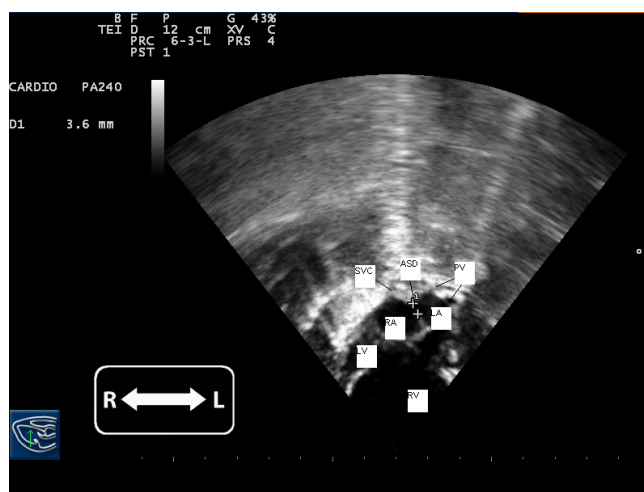


Figure 3. Transthoracic echocardiography subcostal four-chamber views. Note normal pulmonary venous drainage, systemic veins connecting to the right-sided morphologic right atrium, discordant atrioventricular connections and atrial septal defect.

ASD: Atrial Septal Defect, RA: Right Atrium, RV: Right Ventricle, LV: Left Ventricle, PA: Pulmonary Artery, PV: Pulmonary Veins, SVC: Superior Vena Cava

The patient required intubation and mechanical ventilation on admission and during the follow-up; he was extubated after 24 hours. On second week of his admission, oxygen saturations decreased to 60%. Echocardiography showed increase in infun-

dibular and pulmonary valvular gradient. He underwent surgical placement of a palliative systemic-to-pulmonary artery shunt (a modified Blalock-Taussig shunt). His oxygen saturations increased to 90% after the operation.

However, he died two days after the operation because of neonatal sepsis. Autopsy was not performed since his family did not permit.

Discussion

The common variants of double outlet right ventricle are those with the interventricular communication in subaortic position, with the aorta spiralling from right to left relative to the pulmonary trunk, along with pulmonary stenosis, the so-called Fallot variant, also those with the interventricular communication in subpulmonary position, with the aorta to the right of, and parallel to, the pulmonary trunk, the so-called Taussig-Bing variant. Those with the interventricular communication in subaortic position, and with the aorta spiralling from right to left relative to the pulmonary trunk, but in the absence of pulmonary stenosis [2]. Less common variants of double outlet right ventricle are those with the interventricular communication uncommitted, often described as non-committed, to either subarterial outlet, and with the aorta to the right of the pulmonary trunk, with either spiralling or parallel arterial trunks. Those with the interventricular communication in doubly committed position, with the aorta to the right of the pulmonary trunk, and with spiralling arterial trunks. Those with the interventricular communication in subaortic position, but with the aorta to the left of the pulmonary trunk with parallel arterial trunks. Those with mirror imaged atrial arrangement. Those with isomeric atrial appendages and, hence, mixed and biventricular atrioventricular connections. Those with usual atrial arrangement and discordant atrioventricular connections, usually with the aorta parallel to and to the left of the pulmonary trunk. In our patient, interventricular communication was in doubly committed position, aorta was parallel and to the left of the pulmonary trunk with usual atrial arrangement [2].

In the literature, double outlet right ventricle has been described with ventricular inversion form with situs solitus, and with atrioventricular concordance in situs inversus [4-6].

Double outlet right ventricle with discordant atrioventricular connections have been described in association with both usual and mirror-image atrial arrangements. Many patients with usual atrial arrangement have their hearts placed in the right chest, while those with mirror-imaged atria have left-sided hearts. Pulmonary stenosis was present in four-fifths of the largest reported series, with the interventricular communication being subpulmonary more frequently than subaortic. Despite the discordant atrioventricular connections, there was marked variability in the relationships of the great arteries, with the aorta to the left of the pulmonary trunk in only two-thirds of cases. This variant of double outlet right ventricle has much in common with congenitally corrected transposition [2].

Surgical repair of the double outlet right ventricle depends basically on the anatomic type of the condition. Among the choices available for definite surgical repair, the intraventricular repair, connecting the aorta to the morphologically left ventricle through the ventricular septal defect while maintaining the con-

tinuity between the right ventricle and the pulmonary artery, is the preferred operative approach, whenever possible. This technique yields the best results, with less in-hospital morbidity and a lower incidence of reoperation. It is the treatment of choice whenever the ventricular septal defect is subaortic and may also be used in patients with other types of ventricular septal defect [7]. Other forms of definite surgical repair include the anatomical repair (translocation of the great arteries or Jatene's arterial switch procedure), which directs the blood flow from the ventricular septal defect to the pulmonary artery using a surgical flap; the Rastelli repair (which uses a conduit from the right ventricle to the pulmonary artery) and the insertion of a surgical flap directing the blood flow from the ventricular septal defect to the aorta; the Damus-Kaye-Stansel operation, and the inversion at the atrial level, as performed by Senning or Mustard, which directs the flow from the ventricular septal defect to the pulmonary artery [7]. When the ventricular septal defect is subpulmonary, the preferred surgical approach is Jatene's operation; however, this procedure is still a challenge for many surgeons, and the results obtained are less satisfactory than those achieved in patients who undergo the intraventricular repair.

More complex forms of double outlet right ventricle have been successfully repaired. Double outlet right ventricle with atrioventricular discordance can be corrected by closure of the ventricular septal defect, transection of the pulmonary artery, and establishment of morphologic left ventricle to pulmonary artery continuity with an extracardiac conduit. In these patients, the right ventricle remains as the systemic ventricle. Kiser et al. [6] reported repair of dextrocardia, double outlet right ventricle with atrioventricular discordance and ventricular septal defect. More recently, efforts have been directed to establish continuity between the morphologic left ventricle and the ascending aorta through the ventricular septal defect to allow more physiologic function of the left ventricle as the systemic ventricle. However, this also requires a concomitant atrial switch procedure to direct the pulmonary venous blood to the morphologic left ventricle [8].

Fox et al. [9] reported 26 patients with atrioventricular discordance and a variety of ventriculo-arterial connections. Two of them had double outlet right ventricle with atrioventricular discordance. In one of the patients, ventricular septal defect was repaired so that venous blood passed from left ventricle across the defect to the pulmonary artery. The anatomy did not permit this technique in the other patient, therefore, after closing the defect and closing the origin of the pulmonary artery from the right ventricle, a valved external conduit was placed between left ventricle and pulmonary artery. Both of these patients developed permanent atrioventricular dissociation. Both of the patients survived and continue to be well without evident tricuspid valve incompetence [6,9].

Shin'oka et al. [10] reported a total of 189 patients with congenitally corrected transposition of the great arteries or double outlet right ventricle with discordant atrioventricular connections diagnosed between years 1972 and 2005. The median age of the patients was 8.3 years, (range 2 months to 47 years old). 92/189 patients had double outlet right ventricle and atrioventricular discordance. Surgical procedures included 56 Mustard/

Senning–Rastelli procedure, 18 conventional Rastelli, 15 Fontan operations, two double-switch operations, and one conventional technique [10]. In our patient, due to presence hypoplastic pulmonary arteries, surgical placement of palliative systemic-to-pulmonary artery shunt (a modified Blalock-Taussig shunt) was required before definite surgical repair.

We reported a rare complex cardiac malformation; double outlet right ventricle with discordant atrioventricular connections and hypoplastic pulmonary arteries diagnosed by echocardiography. Echocardiography is an effective and extremely useful method for diagnosing this complex malformation as it accurately identifies the anatomical variables and guides the choice of the most appropriate surgical approach through the detection of additional malformations. Segmental approach and sequential analysis of cardiac segments by echocardiography are the clue for diagnosis of complex cardiac malformations.

Competing interests

The authors declare that they have no competing interests.

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